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BY

PARKER SYMS, M.D.

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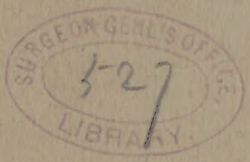

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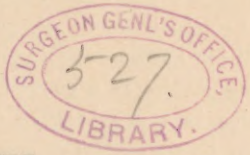


THE ARTHROPATHIES OF LOCOMOTOR ATAXIA.*

By PARKER SYMS, M. D.

In the issue of July 7, 1888, of the *New York Medical Journal* I published a paper on Arthropathia Tabidorum. My excuse for addressing you now on this subject is my desire to emphasize some points of importance, to state the results of my continued study of the disease, and because I believe this theme to be one of particular interest to the general practitioner, and yet one concerning which very little has been written.

The study of structural diseases of joints depending upon central and peripheral nerve lesions is of comparatively recent date. As progress has been made in this direction it has been found that a number of diseases of the nervous system may be responsible for grave arthropathies. These nerve lesions are usually those which belong to the degenerative variety, in which the essential tissues of the nerve centers or trunks are so altered as to no longer be proper nerve tissue, or in which there is absolute destruction or loss of the nerve structure. Among this class those dis-

* Read before the New York State Medical Association, October 11, 1894.

eases which involve the posterior portion of the spinal cord, affecting sensation and circulatory conditions, are the ones which are particularly responsible for the joint diseases. Principal among them are locomotor ataxia and the condition known as syringomyelia.

Syringomyelia is a condition in which there are clefts or cavities in the substance of the spinal cord. These clefts may be congenital or may be the results of degenerative disease. In the latter case the disease consists of three stages: (1) The spinal cord is infiltrated with round cells and connective-tissue nuclei; (2) the replacement of the medullary elements by a feltlike connective-tissue increase or gliomatous mass; (3) this has a tendency to break down in its center, leaving cavities lined with this feltlike connective tissue. These cavities usually contain clear serum, sometimes bloody or hyaline material.

Locomotor ataxia consists pathologically in degeneration of the posterior columns of the spinal cord, apparently extending from the center outward; it finally involves the posterior nerve roots, often the lateral columns, and, as has been shown by careful investigators, the nerve trunks and terminals are also involved. The degeneration consists in atrophy and loss of the essential nerve tissue, accompanied by an increase of connective tissue and a deposit of fat.

That these and similar diseases do cause very grave and remarkable arthropathies is well known, and yet the exact relation of the spinal lesion to the joint trouble does not seem to be clear, and many opposed opinions and theories have been advanced.

The study of the ætiology of the arthropathies of locomotor ataxia needs first the study of the parent disease and subsequently a study of the joint trouble.

The causes of locomotor ataxia are certainly not known

and are only surmised. All that can be said is that it is more common among men than among women; that it is a disease of middle and advanced life. In a very large percentage of cases the patients have had syphilis. In the majority of cases one can obtain a history of excessive venery, of excessive brain and nerve tax, or excess in the use of alcohol. Often the onset is charged to some severe crisis involving exposure to wet and cold combined with physical exhaustion. There has been a great deal of dispute concerning the relation of syphilis to tabes dorsualis. I believe the majority of authors regard syphilis as the cause of locomotor ataxia—*i. e.*, that the disease occurs as the direct result of syphilis, and never in patients who have not been so infected. A few regard a previous history of syphilis as merely a coincidence and not as a cause, while some regard syphilis as one of several predisposing causes of the spinal lesion. My own conviction, based on the study of a sufficient number of joint cases, is that locomotor ataxia may occur independently of syphilis; that any prolonged and destructive tax upon the nervous system may so exhaust it that the circulatory conditions shall become unbalanced and the nutrition of the cells so altered as to result in degeneration of the nerve substance. Excess of venery, of prolonged intense nerve work, prolonged deprivation of sleep, rest, and nutrition, excess in the use of alcohol and in the exercise of nerve energy under stimulation, all diminish the opportunity for physiological rest and for cell repair. Any grave disease, as tuberculosis or syphilis, also taxes the system and not only may, but certainly does, act as a factor in the production of disease of the nerve centers. Certainly, if syphilis is necessary for the production of tabes dorsualis, there can be no case without it; but in a large number of tabetics one can neither obtain a history of syphilis nor can he demonstrate any syphilitic lesion.

Syphilis is unfortunately a very evident disease, and the cases which are severe enough to be the cause of locomotor ataxia may reasonably be expected to leave some impression on the patient's mind, if not upon his body. Certainly it is but surmise to allege that syphilis has been found in a patient when no evidence of the disease exists and none can be recalled. It must be conceded that excess of venery is a predisposing cause of syphilis. Perhaps the excess which made the syphilitic infection possible may also have damaged the nervous system by exhausting and overtaxing its powers of repair.

Concerning the ætiology of the joint lesions in locomotor ataxia, I take the liberty of quoting from my former paper. It has been maintained:

1. That it is purely a mechanical condition caused by the abnormal gait of the patient, due to locomotor ataxia.

2. That it is due to a direct trophic disturbance of central origin — a certain point being involved for a corresponding joint, or that there is one center responsible for the nutrition of joints. (Buzzard suggests that this hypothetical point is in the medulla.)

3. That it is not a peculiar disease, but is (*a*) arthritis deformans coincident with the tabes, or (*b*) a purely syphilitic joint in the same connection.

4. That the serous effusion is due to trophic disturbance, and that the changes in the bone are due entirely to the pressure of the effusion and the consequent anæmia of the periosteum. (Macnamara.)

5. That it is due entirely to a traumatism and that the primary changes are inflammatory, but, owing to analgesia, the joint is not kept at rest and progress is different, merely on that account, from ordinary traumatic arthritis.

6. That it is due to a direct trophic disturbance, like ulcus perforans, not of central origin, but due to atrophy of

the peripheral nerves (Weizsäcker). I understand this to mean that only the nerves of the diseased joint were atrophied, and therefore this joint became involved.

7. That it is due to traumatism or some change in the condition of the joint to be affected. The progress attained depends on the fact that the nerves of all large joints are degenerated, and some degeneration occurs in the structures of apparently healthy joints in tabetics, as pointed out by Jurgens and Westphal.

I am inclined to the last theory. The fact that the nerves and structures of seemingly healthy joints are in a state of partial degeneration has been demonstrated, and it only remains to assign a reason why a certain joint should continue in the process. I think it is reasonable to look for this in some local condition, such as traumatism. If it were simply a continuation of the degenerative process, other joints would be involved in time. But a patient may have a single joint affected for years.

The idea that it is dependent on the peculiar gait is not tenable, for the disease occurs before inco-ordination is present, and patients have developed it while lying in bed; also because it occurs in the upper extremity.

Neurologists have not found for us the central point alleged by Buzzard and Charcot, and changes in the peripheral nerves seem to explain the disease more satisfactorily. Charcot's disease differs in its pathology and symptoms from arthritis deformans and syphilitic joints sufficiently to give it an identity as a peculiar condition.

The idea that it is caused by the serous effusion, as suggested by Macnamara, it seems to me does not need contradiction.

I do not consider it of a purely traumatic nature. The changes are none of them of an inflammatory nature, and certainly must be regarded as due to nerve changes.

The pathological changes in these joints are remarkable, involving both the bone and the soft parts, producing the most extreme deformities. The synovial membrane is usually anæmic, though sometimes congested. The capsule becomes very much thickened by connective-tissue increase. The circumarticular structures are often the seat of extensive connective-tissue hyperplasia, and this mass forms a large part of the deformity. The entire capsule may be destroyed and be replaced by a mass of new connective tissue. There is always a hydrarthrosis early, and a peculiar effusion into the deep fascia and the soft parts, sometimes extending throughout the limb. The capsule and ligaments are stretched and distended by the swelling and hydrops. This gives an abnormal mobility to the joint. Ossification occurs in any of the tissues at and about the joint. Bone formations may be found in the tendons, muscles, fasciæ, and capsules, or they may be formed as free bodies in the joint cavity. There are two classes of cases, owing to two sets of processes which occur in the bone tissue—viz., the hypertrophic and the atrophic. We may find a joint diseased in both manners—that is to say, one bone of the joint may be hypertrophied and the other atrophied. In the atrophic form the ends of the bone and the epiphyses are more or less absorbed, and the bone may terminate in a rounded end like a drumstick.

The hypertrophic form produces great increase in the size of the bone by large bony deposits and outgrowths on the epiphysis. These bony deposits are not confined to the epiphyses, but also involve the shaft, and are often found on the capsular and soft parts. Spontaneous fractures are very common. They may involve the shafts or the processes. These fractures may unite, often forming good union, but the callus is usually overabundant. The bone structure is changed microscopically and chemically. As in osteitis rarificans, the lamellæ are absorbed and broken

down so that several canaliculi are transformed into one large canal. The bone plates are thinned, and the canals are filled with fat. As in osteomalacia, the phosphates are diminished to about eleven per cent., and the fat is increased to about thirty-seven per cent.

In the hypertrophic forms the joint functions are usually better than in the atrophic. In the latter the opposing surfaces are diminished, and the lack of support allows great freedom of abnormal motions.

The symptoms are well marked, and the disease is easily recognized. As these joint affections come early in the course of locomotor ataxia, we may not have many symptoms of that disease present to aid our diagnosis; but we are sure to find some. The prominent symptoms of tabes are darting neuralgic pains, numbness and peculiar sensations in the feet and legs, perverted response to surface irritation, as an exaggerated sensation of cold applied to the skin, or the application of cold may produce a sensation of heat, etc. Tactile sensation is often delayed. Analgesia is a prominent early symptom of tabes, and is a constant condition. So is loss of patellar reflex. The Argyll-Robertson pupil is one of the characteristics of the disease. Inco-ordination of the muscles of the extremities is very prominent as a symptom, but it comes on late in the disease. Owing to the blunted sensation in the feet there is more or less inability on the part of the patient to walk or stand steadily without the aid of sight, and in the dark, or with the eyes closed, he will sway and stagger. Inability to walk at night and the peculiar sense of numbness are usually the first warnings to the patient of this disease of the spinal system.

The joint disease is characterized by its sudden onset and comparative absence of pain. Sometimes the first indication of the disease is a spontaneous fracture of a bone shaft, but usually there is a sudden and rapid swelling of

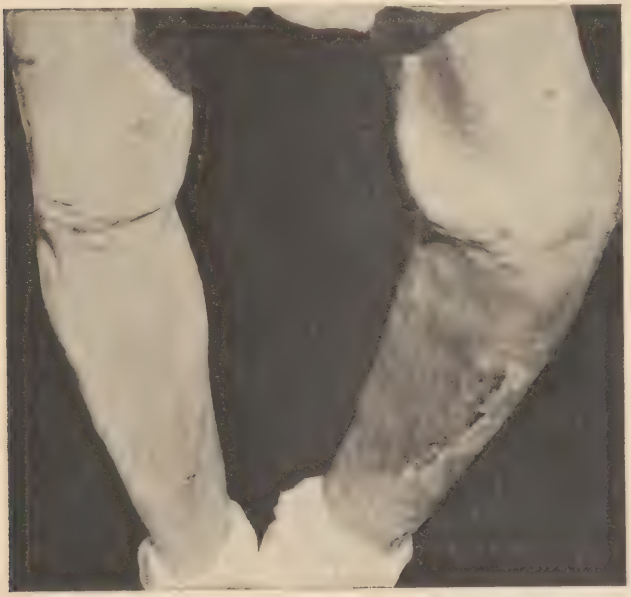
the limb produced by an œdema of the deeper parts. This general swelling subsides, but the joint is found to be deformed, and remains enlarged. The peculiar deformity of the joint, its rapid development, the absence of pain, attended by some of the characteristic symptoms of locomotor ataxia, complete the means of diagnosis. It may be mistaken for arthritis deformans, but this latter disease comes on slowly, is attended by severe pain, and you will be aided in differentiating by the absence of the symptoms of tabes. Besides, in arthritis deformans the motion of the joint is restricted, while in arthropathia tabidorum the joint is usually abnormally movable.

In these cases the disability is great, especially late in the spinal disease when inco-ordination is added; but I have never seen a patient who could not walk with the aid of crutches or a cane. One of my cases has improved in his ability to walk owing to the fact that his knee, the seat of the disease, has become ankylosed, and, though still greatly deformed, it is strong and gives him firm support. Six years ago, when I first saw him, mobility was so great that he walked with much difficulty. Knowing that in these cases fractures would unite, I was of the opinion that resection of the knee in this case would be justifiable, and that he would be much improved by the operation. Nature has accomplished all that I could have hoped to do by operating, and I now know that I might have failed in my object.

This week I saw a case of this disease in a knee joint in which resection had been done seven years ago by Dr. Willy Meyer, of this city. The final result shows that union of the bones had failed to take place; and, while the man can walk with a cane, the joint is not better than the average of these cases in which no operation has been done. As I am ignorant of the man's original condition I am unable to state whether the operation was an advantage or not.

Beyond the use of splints in those cases where mobility is great, very little can be done in the way of treatment. The disease is permanent, and the patient I have referred to is the only one of my acquaintance who has improved. Further experience may show that resection is advisable. Notwithstanding the case above referred to, I am still of the opinion that we may expect union to take place after resection. If this is true, the operation would often afford great relief.

I have had no reason to change my conclusions as ex-



Case I.

pressed six years ago. They are as follows: (1) That this is a peculiar disease depending on tabes dorsualis; (2) that it is due to trophic disturbance; (3) that it is of

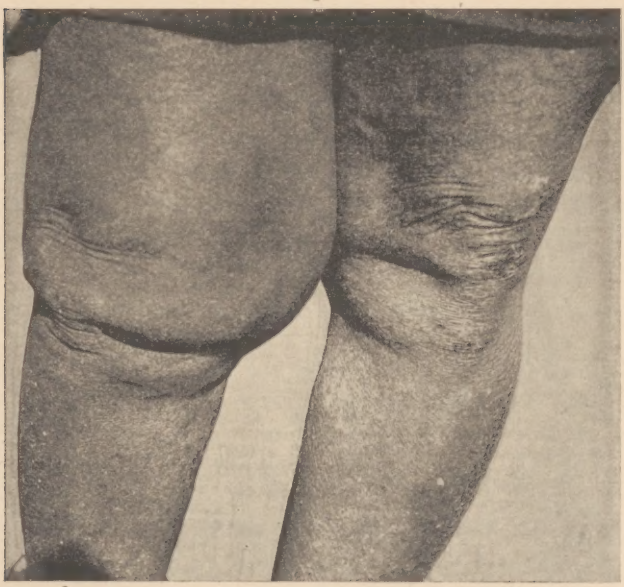
the nature of a degeneration, and not of an inflammation; (4) that syphilis is not a necessary factor of locomotor ataxia; (5) that in some cases excision of the diseased joint may be justifiable.

The following is the history of two typical cases which I take pleasure in showing you:

Case I.—Henry B., sixty-two years of age, a sea cook by occupation. No history of syphilis and no evidence of syphilis. When six years of age was kicked by a horse, probably receiving a fracture of the right parietal bone. He says he was unconscious for six days, and that he bled a good deal; does not know if from ear or nose. There was no resulting paralysis of speech or motion. Since this accident he has been deaf in the right ear. His subsequent history is negative till 1883. At that time, after taking a bath and exposing himself to cold, he suddenly developed what he calls a weakness in his legs. His gait was unsteady, and he was thought to be intoxicated. Soon he lost to a marked degree the natural sensation in his legs and feet; they felt numb and as though he walked on an uneven, spongy surface. His symptoms have always been more marked on the left than on the right side. There was very little change in his condition till about the middle of the year 1887. At that time his knee swelled to more than its present size within a week. It attained its present deformity within a month. The joint was very movable in all directions. When he stood it took a position of extreme adduction. When I first saw him, in the latter part of 1887, there was a peculiar hard swelling of the entire limb. There was a large, imperfectly movable mass below the middle of the thigh. This later became reduced in size, and ossified on the shaft of the femur. Examination five months after the attack of general swelling of the thigh showed anæsthesia, deep and superficial, of both limbs; sensation not delayed; patellar reflex absent; pupils do not respond to light; patient sways when standing or walking with closed eyes; left knee very much enlarged and deformed, measuring ten centimetres more than the right knee. There was some increase in the size of the entire limb; there was fluid in the joint cavity, marked crepitus on motion; lateral

mobility greatly increased; general contour of joint lost, owing to thickening of the capsule; internal capsule greatly hypertrophied. There was also a spine of bone, ten centimetres long, on the inner side of the lower end of the shaft of the femur.

Examination on October 8, 1894, showed the left knee joint very much enlarged and deformed, measuring ten centimetres more than the right knee, over the patella; the knee was ankylosed; the position of leg was not one of normal extension, but adduction, causing a marked bow. Owing to the ankylosis, the limb gave a firm support, and the patient was able to walk quite well.



Case II.

Case II.—Laura H., admitted to the Colored Hospital on March 21, 1894; fifty-four years of age. Evidence of old syphilis, but no history of the same. The present illness began in the early part of 1892, with numbness in both feet. In the

fall of 1892 she began to have shooting pains in calves of legs. About December, 1892, her right knee became swollen and caused her some pain. There were present the Argyll-Robertson pupil and loss of patellar reflex; sensation in the legs was delayed, and was also diminished; she swayed when standing with closed eyes. The right knee was greatly deformed and enlarged; the patella was firmly fixed to the femur; the lower end of the femur was greatly hypertrophied, especially at the inner condyle. The ligaments and capsule were much distended and stretched. The upper end of the tibia was somewhat atrophied, and was subluxated backward and outward. There was flexion to more than ninety degrees; extension to more than a straight line; abduction to about a right angle; adduction to about forty-five degrees. The right knee measured seven centimetres more than the left at the greatest circumference. The disability was great.

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FRANK P. FOSTER, M.D.

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